Sudden Headaches

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Abstract

Sudden severe headache is a cardinal feature of subarachnoid haemorrhage (SAH) and other potentially life threatening neurological conditions. Early diagnosis and management improves the outcome of SAH. In general practice, benign thunderclap headache vastly outnumbers SAH as a cause of sudden headache. Thunderclap headache is a diagnosis of exclusion and all patients admitted with a first presentation of a severe headache lasting more than one hour should be investigated with a CT and lumbar puncture. It must be borne in mind that these investigations may miss diagnoses such as venous sinus thrombosis and Magnetic Resonance Angiography (MRA) should be performed if clinical suspicion remains high.

Key Words

Subarachnoid haemorrhage, thunderclap headache, lumbar puncture.

Introduction

Patients with headache account for 1–2% of attendances in Accident and Emergency.[1] As many as a third of patients presenting to A and E with acute headache will be harbouring potentially fatal or disabling intracranial conditions such as subarachnoid haemorrhage (SAH). This article is written primarily as a guide for the general medical trainee when faced with a patient with a sudden headache, focusing on the diagnosis and early management. The primary objectives are to make the diagnosis, relieve the headache, investigate and manage appropriately. Many patients will be in distress and there is a tendency to channel the patient immediately towards the nearest scanner. However, the importance of a careful history should not be overlooked.[2]

Subarachnoid haemorrhage

The case fatality rate of SAH is around 50% overall and one-third of survivors remain dependent. It is important to remember that early recognition and treatment of SAH may improve outcome and in the UK, failed diagnosis or management of SAH represents the largest single area of neurological litigation. On the other hand, the low incidence of SAH (around 6 cases per 100,000 patient years in the UK) and high frequency of headache in general practice might result in patients being referred for investigations unnecessarily.

Cause of SAH

About 85% of all spontaneous SAHs result from rupture of a saccular aneurysm at the base of the brain. The prevalence of unruptured aneurysms, as derived from prospective autopsy series, is in the region of 3/100 patients. This means that the vast majority of aneurysms never rupture.[5] The cause of aneurysm development is unknown, although smoking, hypertension and alcohol abuse are all risk factors for SAH. Rarely, aneurysms develop from septic emboli (for example in patients with infective endocarditis) or from cocaine abuse.

Of the remaining 15%, a few are caused by rare disorders such as dural arterio-venous malformation and arterial dissection. About 10% are caused by perimesencephalic haemorrhage. In this radiologically distinct and strikingly harmless variety of SAH blood is confined to the cisterns around the midbrain. Loss of consciousness, seizures and focal signs are rare but there is otherwise little to distinguish the headache clinically from aneurysmal SAH.

Clinical features

The clinical hallmark of SAH is an unusually severe headache that started suddenly. Many will have features of meningism (neck stiffness, photophobia and Kernig's sign) due to irritation of the meninges by free blood in the CSF. Almost half of patients are unresponsive for more than an hour and one third have focal neurological signs which develop at the same time as the headache or soon after.[3] These patients should not pose any dilemma in whether or not to investigate further.

In those whose only symptom is headache it is often difficult to recognise the seriousness of the underlying condition. Even an accurate history may not reliably distinguish SAH from more benign headaches.[4] The following statements are worthy of note:

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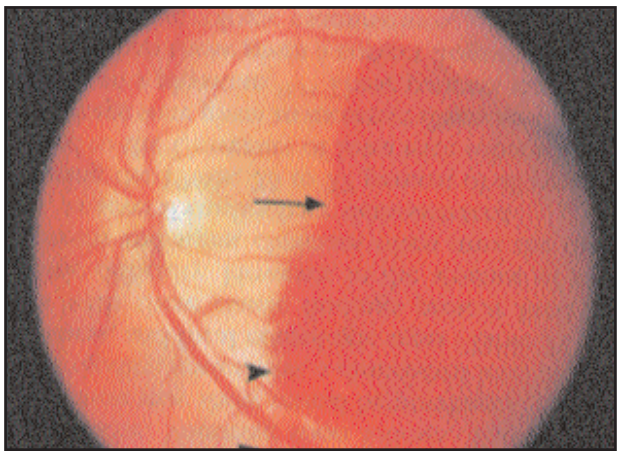

Figure 1. Subhyaloid haemorrage. Arrows indicate margin of haemorrage

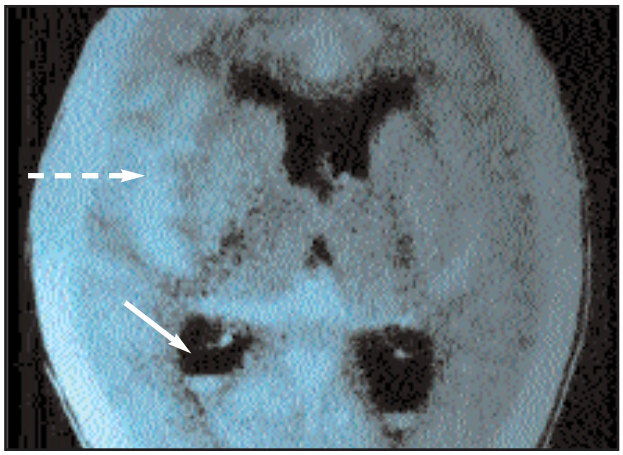

Figure 2. CT head scan without contrast showing acute haemorrage into the ventricles, subarachnoid haemorrage (solid arrow) and parenchyma, intracerebral haemorrage (dashed arrow)

1. Only half of patients with SAH describe an instantaneous headache, the reminder describe onset over several minutes.
2. In those with instantaneous headache, innocuous headache outnumbers SAH by 10 to 1 in the general population.
3. The severity of the headache is in those with SAH and benign thunderclap headaches is rated equally.
4. Vomiting occurs in 70% of patients with SAH but also 43% of those with benign thunderclap headache.
5. Preceding bouts of 'sentinel' headache are recalled in 20% of patients with SAH and in 15% of patients with benign thunderclap headache.
6. Neck stiffness is common in SAH, but may develop later.
7. The classic sign of SAH, subhyaloid haemorrhage (figure 1), is most common in patients with increased intracranial pressure and a decreased level of consciousness.

It is even more difficult to suspect a SAH if there is no history of a sudden headache or if other symptoms such as seizures (presenting feature in 6-16% of SAH) or confusion (presenting feature in 1-2% of SAH) prevail. Trauma and SAH may be difficult to disentangle, for instance in those who have sustained a skull fracture after aneurysm rupture, when head trauma caused an aneurysm to burst.

The headache of SAH usually lasts from days to weeks, but may be of shorter duration. Most specialists agree that SAH is unlikely if the headache lasts less than one hour. However, the shortest duration of headache associated with SAH is unknown, and SAH should be still be considered possible in sudden severe headache of any duration, particularly if there is associated impairment of consciousness. The presence of nausea, vomiting, neck stiffness, transient loss of consciousness, or focal neurological symptoms are supportive of the diagnosis of SAH, but are not sufficient to distinguish from more benign syndromes.[4] Absence of these symptoms cannot be taken as reassurance that the patient has not had an SAH, as a proportion will present with headache as the only symptom.

Investigations

CT Scanning

All patients with a suspected SAH should have an immediate CT head scan. The ability to detect subarachnoid blood declines progressively from the onset of symptoms, so that early scanning is crucial. The pick-up rate is also critically dependent on the experience of the observer. (figure 2)

Lumbar puncture

However early the CT is performed some SAH will go undetected. All patients who have had a clinical suspicion of a SAH high enough to merit a CT scan should undergo lumbar puncture (LP) if the CT scan is normal, to look for red blood cells and xanthochromasia.

The 3 tube test (i.e. a decreasing CSF red blood cell count in 3 consecutive tubes being indicative of a bloody tap from a haemorrhage) is unreliable. Only the absence of xanthochromasia, formally assessed by spectrophotometry, can be relied on to exclude SAH following LP.

Xanthochromasia refers to the yellowish discolouration of CSF supernatant from breakdown products of haemoglobin: oxyhaemoglobin and bilirubin. After a SAH red cells are gradually lysed within the CSF. Released haemoglobin is metabolised to oxyhaemoglobin and bilirubin. Oxyhaemoglobin can be detected within hours, but bilirubin requires up to 12 hours to appear. Timing of a LP is therefore crucial and should be delayed until at least 12 hours after the onset of the symptoms.[6] The CSF should be centrifuged and examined promptly so that red cells from a bloody tap do not undergo lysis in vitro. Xanthochromasia is found in all SAH from 12 hours to 2 weeks after the haemorrhage but declines thereafter (Table I).

CT – detection of SAH		LP – presence of bilirubin	
<24 hours	98%	1-12 hours	variable
24-48 hours	86%	12 hrs- 2 weeks	100%
2-5 days	76%	3 weeks	70%
5 days	58%	4 weeks	40%

Table I. Reliability of CT and lumbar puncture in detection of subarachnoid haemorrage and time intervals after onset of headache

Cerebral Angiography

The advent of new imaging techniques such as CT angiography and MRA have reduced the need for cerebral angiography, but it remains the gold standard investigation to identify and treat a bleeding aneurysm. The technique carries about a 1% risk of mortality and morbidity and so should be reserved for those with confirmed SAH. Usually a '4 vessel' angiogram (i.e. both carotid and both vertebral arteries) is performed to pick up multiple aneurysms on other vessels.

Early complications of SAH

All patients should observed closely. Up to 15% of patients have an acute deterioration in the first few hours after admission, usually due to rebleeding. Other patients may become obtunded due to development of a haematoma requiring urgent surgical evacuation. Hydrocephalus (CSF flow obstructed by blood) presents with gradual obtundation over 24 hours and unless urgently treated with neurosurgery has a very poor outcome.

Treatment of aneurysmal SAH

Once a diagnosis of SAH has been established, or in a suspected case with a deteriorating level of consciousness, the patient should be discussed urgently with the local neurosurgical unit.

The primary aim in management of SAH is prevention of secondary cerebral ischaemia due to vasospasm. Therapies include:

- Careful fluid management is to prevent a reduction in plasma volume which may contribute to cerebral ischaemia. A daily intake of at least 3 litres of saline has been shown to reduce cerebral ischaemia and improve overall outcome.
- Nimodipine, a calcium antagonist, improves outcome in SAH but it is uncertain whether nimodipine acts through neuroprotection, reducing vasospasm or both. The standard treatment is nimodipine 60mg orally (or via NG tube) every 4 hours, continued for 3 weeks.

Other aspects of general management should include:

- Pain control. Start with paracetamol or codeine phosphate. Avoid aspirin.
- DVT prophylaxis. Use compression stockings before occlusion of the aneurysm.
- Laxatives. Constipation from analgesics and immobility is common.

Key points 1

1. No feature in the history (or examination) reliably distinguishes SAH from benign headaches.
2. All patients with a sudden or rapid (minutes) onset of headache should be investigated with a CT.
3. All patients with a normal CT should have a LP, 12 hours after presentation.
4. CSF should be examined for bilirubin by spectrophotometry.
5. 10% of SAH are due to perimesencephalic haemorrhages: no aneurysm can be detected on the angiogram.
6. Incidental, unruptured aneurysms are common in the general population and when small have a low risk of rupture.

Definitive management

Aneurysms detected on angiography may be treated either by endovascular coiling of the aneurysm with platinum wire coils or by craniotomy and clipping of the neck of the aneurysm. The choice depends on individual expertise, the size, position and shape of the aneurysm. Coiling has become increasingly popular due to its lower morbidity compared to craniotomy.

Benign Thunderclap headache

For the vast majority of patients presenting with sudden onset headache, the diagnosis of SAH is excluded by investigations. In such cases the most likely explanation for the symptoms is benign thunderclap headache.

Clinical Features

The headache typically reaches a maximum intensity within 30 seconds and lasts for several hours, but may persist at lower intensity for several weeks. Episodes may occur repeatedly for 1-2 weeks and in up to one third may recur over subsequent months to years. They may occur at rest or may be precipitated by intense exertion, Valsalva manoeuvres or during sexual intercourse (see below).

Pathophysiology

The pathophysiology of idiopathic thunderclap headache is unclear but it may be due to self-limiting diffuse segmental vasospasm. In these cases it may be associated with fluctuating focal neurological signs, seizures or even stroke.[12]

Thunderclap headache may be a migraine variant, since half of patients will have a pre-existing history of, or will subsequently develop, migraine.

Prognosis

Idiopathic thunderclap headache appears to be a benign phenomenon: when 93 patients with sudden headache and negative investigations were followed up for 1 year, none subsequently had SAH, stroke or sudden death.[7]

Idiopathic thunderclap headache is a diagnosis of exclusion; the diagnosis should only be made after negative CT and LP have excluded the possibility of SAH.

Sudden Headaches

Other causes of sudden headache

In one series, 18 of 148 patients presenting with sudden severe headache were found to have other intracranial pathology where SAH was excluded.[7] Causes included meningitis/encephalitis, intracerebral haematoma, AVM, subdural haematoma, giant aneurysm without haemorrhage and haemorrhage into a tumour and middle cerebral artery infarction. The likelihood of serious intracranial pathology in these cases will usually be apparent due to accompanying neurological symptoms and signs. When examination is unremarkable, the following conditions should be considered.

Cerebral venous sinus thrombosis (CVST)

Headache is the most common symptom of CVST and the most frequent presenting symptom in 75% of cases. Although onset of the headache is usually over several days, up to 10% of patients describe a sudden onset. The diagnosis can easily be missed. CT scan is interpreted as normal in about 25% of patients with CVST and up to 40% of patients may have a raised CSF opening pressure without the typical lymphocytic pleocytosis, red cells and elevated protein. Where there is a high clinical index of suspicion (coagulopathy risk factors, oral contraceptive pill, young females) the investigation of choice is Magnetic Resonance Venography (MRV).

Pituitary apoplexy

Pituitary apoplexy results from infarction or haemorrhage into the pituitary gland, usually when it contains an adenoma. Presentation with sudden headache is well recognised. Examination may reveal hypotension, bitemporal hemianopia, ophthalmoplegia and altered mental state, and the CSF may show elevated white and red cell counts. However clinical examination, CT and CSF may also be normal. Pituitary tumours are isodense to brain tissue and can easily be overlooked on CT, even when haemorrhage is present, but are readily identified by MRI (figure 3).

Carotid artery dissection

Headache affects 75% of patients with internal carotid artery dissection, with 13% of patients describing a sudden onset. Pointers to the diagnosis are a hemicranial (frontal) headache and ipsilateral Horner's syndrome due to disruption of sympathetic fibres travelling in the wall of the carotid artery. Unless the patient has already had an ischaemic event, CT and CSF are usually normal and the investigation of choice is MRA. Although no controlled clinical trials are available, prompt anticoagulation with heparin may prevent more serious embolic cerebral ischaemic events.

Coital cephalgia

Benign sex headache, orgasmic cephalgia or coital cephalgia is a well recognised form of thunderclap headache ocurring at the point of climax. It can occur in females as well as males and may be related to the degree of exertion during sex. Like all thunderclap headaches a first presentation should always be investigated thoroughly to exclude a SAH and only when a recurring pattern is established should a diagnosis of coital cephalgia be reached. Prophylaxis with propranolol, indomethacin and calcium antagonists may be helpful.

Key points 2

1. SAH is not the only potentially serious cause of a sudden headache.
2. A normal CT and LP do not exclude serious pathology.
3. MRI and MRA should be considered in those with a high index of suspicion for other pathologies.

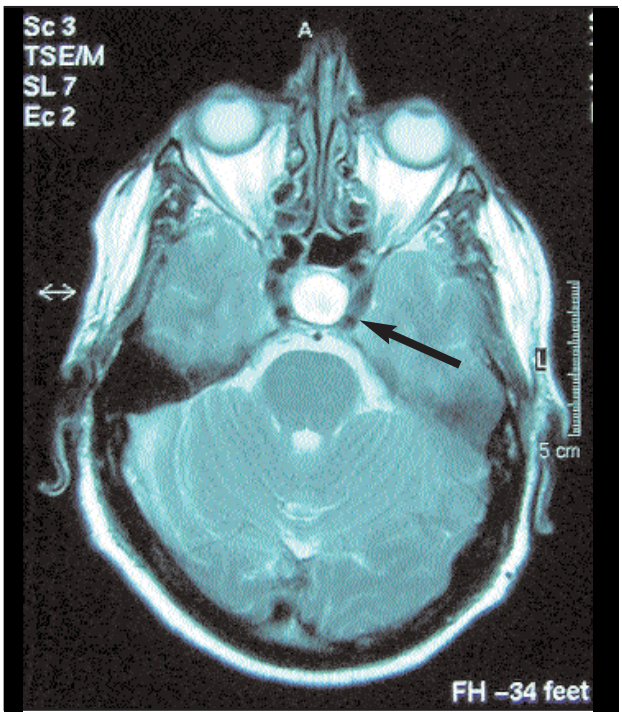

Figure 3. MRI showing a pituitary mass in a 70 year old male who presented with a sudden headache vomiting and dizziness. The CT was reported as normal, but in retrospect showed an enlarged pituitary fossa. He had profound hyoppituitarism

Conclusion

All unusually severe and acute headaches lasting more than 1 hour, should be investigated thoroughly with a CT and LP, proceeding to an MRA if the clinical suspicion of a secondary headache remains high.

Serious neurological disorders may be found in up to 37% of patients, including SAH in 25%.

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